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HEARING LOSS (IN NONOPERATED EARS) IN RELATION TO AGE IN OSTEOGENESIS IMPERFECTA TYPE I

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Hearing loss was studied in relation to age in nonoperated ears in a group of 142 subjects with autosomal dominant osteogenesis imperfecta type I, which was compared to that in a random subsample of 70 subjects. In the n = 142 group, particularly below the age of 30 years, considerable selection (ie, for ear surgery) had occurred on hearing loss. The hearing threshold increased gradually with age. A hearing loss of greater than 30 dB (Fletcher index) was observed for 51% of the subjects older than 20 years and younger than 60 years. The median hearing loss progressed from the 10th to the 45th years of life with an average annual threshold increase (ATI) of 1 dB/y (0.5 to 4 kHz) up to 1.7 dB/y (8 kHz). Sensorineural loss accounted for 0.6 dB/y ATI at 0.5 to 4 kHz and 1.3 dB/y ATI at 8 kHz; conductive loss accounted for 0.4 dB/y ATI at all frequencies.

KEY WORDS — air-bone gap, air conduction, autosomal dominance, bone conduction, sensorineural hearing loss, socially adequate hearing.

INTRODUCTION

Hearing loss has been repeatedly reported as a major symptom of osteogenesis imperfecta (OI).[1-8] A variety of definitions of hearing loss and examination methods can be found in these reports.[5,7-12] The groups of affected subjects examined do not usually represent random samples.

Despite the frequently described progression of hearing loss with age, age has seldom been taken into account in the analyses of hearing loss caused by OI. Obviously, the progression and the extent of hearing loss depend heavily on the age of the affected subjects. The type of hearing loss may be conductive, sensorineural, or mixed.

The present study analyzes the type and extent of hearing loss (in nonoperated ears only) in relation to age in a selected sample of 142 subjects with the autosomal dominant type I of OI.[13] The results are compared to those of a previous study on a random sample of 70 affected subjects, who also had OI type I, from fully examined sibships.[8]

SUBJECTS AND METHODS

Subjects. We were able to collect a group of 142 subjects affected with OI type I for this study. This group is designated here "the selected sample," because either the patients had been previously elected for ear surgery or they volunteered for this study because of their hearing loss. The clinical diagnosis had already been established elsewhere, based on the medical history and physical examination (ie, the presence of blue sclerae, fractures, hyperlaxity, and other possible characteristics in the proband and at least 2 generations of the family). Most of the patients were personally examined, except for 25 patients who had their clinical examination elsewhere at our request.[8] Otorhinolaryngological examination had thus been performed in all of the subjects. Our previous reports concerned the findings and the results of ear surgery in subsamples of the present series.[7,14,15] A segregation and penetrance analysis of hearing loss in a random subsample of 70 affected subjects had been established by fully examining the sibships in 30 families with at least 1 affected subject in 2 generations with elimination of the probands.[8] Only 60 of these 70 patients had undergone clinical audiometry, 2 of them for 1 ear only. Of the other 10 patients, 2 were deceased, 6 were too young for audiometry, and 2 were living abroad.

Experimental Procedure. Pure tone thresholds in decibels hearing level (HL) were obtained from nonoperated ears under standard conditions in a sound-treated room at our clinic or elsewhere, according to ISO 8253[16]; calibration was performed according to ISO 389[17] and ISO 7566.[18] Subjects younger than 4 years of age were excluded from this study for practical reasons. Also excluded were subjects with hearing loss that could be attributed to causes other than OI, such as chronic otitis media. When several audio-

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TABLE 1. SURVEY OF STUDIES ON HEARING LOSS IN OI

Authors	Year	No. of Pts	Limited to OI Type I	Definition of Hearing Loss*	Clinical Audiometry	Hearing Loss Related to Age	Random Sample	Proportion of Patients With Hearing Loss	
								No.	%
Bell[19]	1928	346	–	–	–	–	–	228/346	66
Stoller[1]	1962	26	+	–	Unknown	–	1 family	11/26	42
Carey et al[2]	1968	23	+	–	Unknown	–	1 family	10/23	43
Dufour and Bertrand[20]	1972	11	+	–	Unknown	–	1 family	4/11	36
Carruth et al[21]	1978	10	+	–	+	+	– (3 families)	5/10	50
Quisling et al[4]	1979	68	+	–	+ and portable	–	– (1 family)	20/42	48
Riedner et al[9]	1980	70	–	+	+ and portable	+ (in decades)	– (13 families)	29/70	41
Cox and Simmons[10]	1982	30	–	+	Only portable	+ (in decades)	– (5 families)	11/30	37
Shapiro et al[11]	1982	55	–	+	+	+ (±30 y)	–	35/55	64
Pedersen[5]	1984	173	–	+	+ and portable	+ (in decades)	– (75% of Danish population)	88/173	51
Stewart and O'Reilly[12]	1989	56	–	+	+ and portable	+ (in decades)	–	31/53	58
Garretsen and Cremers[8]	1991	70	+	+	+	+ (in decades)	+	30/70	43
This study		142	+	+	+	+ (in decades)	–	111/142	78

OI — osteogenesis imperfecta, portable — portable equipment implies less accuracy than usual.
*Definitions of hearing loss may differ among studies. + — present or applicable; – — absent or not applicable.

grams were available that had been performed at different ages, we took the one obtained at the youngest age. We used the following working definitions for the various types of hearing loss, paralleling the definitions given by Shapiro et al[11] and Pedersen.[5]

1. Conductive loss was defined as an average air-bone gap for the frequencies 0.5, 1, and 2 kHz, or else an average air-bone gap at 4 and 8 kHz, that was over 15 dB, and a corresponding bone conduction threshold of less than 15 dB.

2. Sensorineural hearing loss (SNHL) was defined as an average air conduction threshold that was at least 15 dB at the frequencies indicated above; the corresponding air-bone gap was less than 15 dB.

3. A mixed loss was defined as an average air-bone gap of at least 15 dB at the frequencies indicated above; the corresponding bone conduction threshold was at least 15 dB.

The synopsis on hearing loss and age in Tables 1[1,2,4,5,8-12,19-21] and 2[5,8-12,21] provides a comparison to the relevant literature. The different types of hearing loss are shown in two age classes (Table 3[11]). Detailed cross-tables (hearing loss by age) were composed for the present group of 142 subjects and the previously reported random subsample of 70 subjects belonging to the fully examined sibships who were not selected for their hearing loss,[8] in order to make a direct comparison (Tables 4 and 5). Hearing loss was substantiated by comparison with the corresponding 95th percentile value for presbycusis.[22]

The statistical test used was the χ^2 test for a 2×2 table, with a level of significance of $p = .05$ (χ^2 value 3.84). Scatter plots were prepared for the data on hearing loss and age (Figs 1-3); details about the type of threshold data and the analyses involved are given where relevant.

RESULTS

The proportion of affected subjects having hearing loss and the relevant data from the literature are presented in Table 1. The present study shows the highest percentage (78%) of hearing loss reported so far. The exact application of the definitions of hearing loss given by Pedersen[5] and Shapiro et al[11] yielded 80% and 83% hearing loss, respectively, for the present series; this finding indicates that there are only slight differences between the definitions. The

TABLE 2. SURVEY OF STUDIES ON HEARING LOSS IN OI AS DEPENDENT ON AGE

Authors	*Year*	*Definition of Hearing Loss*	*Number of OI Pts*	*Mean Age (Range)*	*4-9 y*		*10-19 y*		*20-29 y*		*<30 y*		*30-39 y*		*40-49 y*		*50-59 y*		*≥60 y*		*≥30 y*	
					No.	*%*	*No.*	*%*	*No.*	*%*	*No.*	*%*	*No.*	*%*	*No.*	*%*	*No.*	*%*	*No.*	*%*	*No.*	*%*
Carruth et al[21]	1978	–	10	44 (8-77)	0/1		0/1				0/2		1/3		0/1		1/1		3/3		5/8	62
Riedner et al[9]	1980	+	70	?	0/10	0	3/8	38	6/19	32	9/37	24	2/9	22	4/8	50	7/9	78	7/7	100	20/33	61
Cox and Simmons[10]	1982	+	30	20 (4-67)	1/8	13	4/10	40	2/5	40	7/23	30	1/3		2/3				1/1		4/7	57
*Shapiro et al[11]	1982	+	55	? (2-64)							18/37	49									17/18	94
†Pedersen[5]	1984	+	173	?	5/23	22	10/36	28	18/31	58	33/90	37	15/29	52	12/21	57	8/12	67	20/21	95	55/83	66
Stewart and O'Reilly[12]	1989	+	53	? (10-60)			2/13	15	6/14	43	8/27	30	5/7	71	11/12	92	7/7	100			23/26	88
Garretsen and Cremers[8]	1991	+	70	? (4-66)	2/18	11	13/21	62	6/13	46	21/52	40	1/8	13	2/3		3/4		3/3		9/18	50
This study		+	142	27 (4-87)	4/13	31	24/38	63	26/31	84	54/82	66	23/24	96	17/18	94	9/10	90	8/8	100	57/60	95

Proportion of patients with hearing loss within given decade is presented. Percentages are given only for n > 4.

+ — present or applicable, – — absent or not applicable.

*Original classes 1st to 3rd decades and 4th to 7th decades.

†Estimated from illustrations in original paper.

random subsample reported by Garretsen and Cremers[8] yielded a much lower percentage (43%) of hearing loss.

It is remarkable that the family studies[1,2,4,9,10,20,21] yielded a similar low percentage (90/212 or 42%). The latter studies, including the work by Garretsen and Cremers,[8] indicate a mean percentage of 43% for hearing loss, which has a corresponding proportion of 120 to 282. This differs significantly from the present and other nonfamily studies[5,11,12,19] for which the percentage of hearing loss is 64%, with a corresponding proportion of 493 to 769.

Table 2 shows the relation between the proportion of subjects with hearing loss among the affected subjects and the age of the subjects (in decades) for the present study and some other relevant studies.[5,8-12,21] For those below 30 years of age, hearing loss was observed for 96 of 268, or 36%. The present study, however, had a significantly higher proportion of subjects with hearing loss (54/82 or 66%). For those age 30 years and over, the present study and the one by Shapiro et al[11] had higher proportions of hearing loss (57/60 or 95% and 17/18 or 94%, respectively) than the other selected groups of patients,[5,12] which showed an average proportion of 78 to 109 (72%). The proportions corresponding with 94% and 95% are significantly (see Subjects and Methods) higher than those for all the family studies[8-10,21] taken together, which yielded a proportion of subjects with a hearing loss of only 38 of 66 (58%). The data in Table 2 clearly show that the proportion of subjects with hearing loss increases with age. This is particularly clear in the larger series. An exception is the series published by Garretsen and Cremers,[8] which showed large deviations from the general trend in the second and fourth decades. Together with the present series, of which it represents a subsample, the series by Garretsen and Cremers differs significantly from the others in the high proportion of hearing loss in affected subjects in their second decade of life.

TABLE 3. TYPES OF HEARING LOSS IN OI TYPE I (SELECTED SAMPLE)

	<30 y		*≥30 y*		*Total*	
	No.	*%*	*No.*	*%*	*No.*	*%*
Normal hearing	52	34	8	6	60	21
Conductive hearing loss	3	2	0	0	3	1
Mixed hearing loss	56	37	89	68	145	51
Sensorineural hearing loss	27	18	25	19	52	18
CHOI*	10	7	7	5	17	6
Deafness	3	2	2	2	5	2
Total	151	100	131	100	282†	100

Number of ears in 142 patients with OI type I with normal hearing or specified type of hearing loss in selected sample (282 ears) in total and in two age classes.

*Hearing loss "characteristic of OI" according to Shapiro et al.[11]

†Threshold unknown in 1 ear in 2 cases.

The statistics for normal hearing and the various types of hearing loss are presented in Table 3,[11] including two age classes. Normal hearing was most frequent at a younger age. Conductive loss was found only in a few young affected subjects. Mixed hearing

TABLE 4. PROPORTION OF HEARING LOSS WITHIN GIVEN THRESHOLD RANGE BY DECADE IN OI TYPE I (SELECTED SAMPLE)

Hearing Loss (dB)	*4-9 y*		*10-19 y*		*20-29 y*		*30-39 y*		*40-49 y*		*50-59 y*		*≥60 y*	
	No.	*%*	*No.*	*%*	*No.*	*%*	*No.*	*%*	*No.*	*%*	*No.*	*%*	*No.*	*%*
0-10	21/26	81	36/76	47	16/60	27	6/50	12	10/36	28	5/20	25		
15-30	5/26	19	14/76	18	14/60	23	12/50	24	9/36	25	8/20	40	3/16	19
35-50			16/76	21	21/60	35	12/50	24	5/36	14	1/20	5	7/16	44
55-90			8/76	11	6/60	10	17/50	34	9/36	25	3/20	15	5/16	31
95			1/76	1	3/60	5	2/50	4	3/36	8	3/20	15	1/16	6
>30	0/26	0	25/76	33	30/60	50	31/50	62	17/36	47	7/20	35	13/16	81

Hearing loss in decibels hearing level (Fletcher index). Shading indicates median hearing loss for each decade.

loss was the most frequent type of hearing loss, and the frequency increased significantly with age. Constant proportions, independent of age, were found for SNHL; for the type of SNHL occurring predominantly or exclusively at 8 kHz, which was considered by Shapiro et al to be "characteristic of OI"[11(p2122)] (CHOI); and for deafness.

Table 4 presents the proportions of patients with a given hearing loss by age (in decades) for the present series of selected subjects. Table 5 is a similar table for the random subsample.[8] In Tables 4 and 5, the median hearing loss per decade is also presented. In the present selected sample, the median hearing loss increased from the first decade, at 0 to 10 dB, up to about 30 to 35 dB in the third decade (Table 4). In the random subsample (Table 5), the median hearing loss only increased after the fourth decade. In the selected sample from the 20th year of life, the air conduction threshold was higher than 30 dB for 51% (85/166) of the affected subjects 20 to 59 years of age (Table 4); a further (significant) increase in this proportion could only be noted in the seventh decade. Testing the proportions in the cells for 0 to 10 dB and the second or third decade in the random sample (Table 5) against the corresponding proportions in the selected sample (Table 4) showed that the random sample had significantly higher proportions in these cells (58% as compared to 47%, and 70% compared to 27%, respectively; see Tables 4 and 5). The difference in the relation between the air conduction threshold and age between the two series (random and selected cases) appeared to be significant. With increasing age, the two series tended to develop a similar (median) hearing loss, but the series of selected cases showed selection, especially at a younger age. For each pair of corresponding cells in Tables 4 and 5, we calculated the difference in proportion between the random and the selected series. In this way we obtained a difference distribution that showed a higher proportion than the random sample (Table 5) in the following cells: 35 to 50 dB for 10 to 19 years (9/24 or 38%); 55 to 90 dB for 10 to 19 years (6/24 or 25%); and 35 to 50 dB for 20 to 29 years (19/40 or 48%). For the three cells combined, the distribution of the difference indicated a proportion of 34/64 (53%), while this combination for the random sample (Table 5) yielded a proportion of only 11/72 (15%); the difference is significant. The difference distribution (not shown) had a median threshold for all age classes beyond the first decade that coincided almost invariably with the class 35 to 50 dB.

In order to establish further characteristics of the

TABLE 5. PROPORTION OF HEARING LOSS WITHIN GIVEN THRESHOLD RANGE BY DECADE IN OI TYPE I (RANDOM SAMPLE)

Hearing Loss (dB)	*4-9 y*		*10-19 y*		*20-29 y*		*30-39 y*		*40-49 y*		*50-59 y*		*≥60 y*	
	No.	*%*	*No.*	*%*	*No.*	*%*	*No.*	*%*	*No.*	*%*	*No.*	*%*	*No.*	*%*
0-10	20/24	83	30/52	58	14/20	70	2/2		2/6	33	2/8	25		
15-30	4/24	17	12/52	23	4/20	20			4/6	67	5/8	63	1/6	17
35-50			7/52	13	2/20	10							4/6	67
55-90			2/52	4							1/8	13	1/6	17
95			1/52	2										

Hearing loss in decibels hearing level (Fletcher index). Percentages are only for n > 4. Shading indicates median hearing loss for each decade.

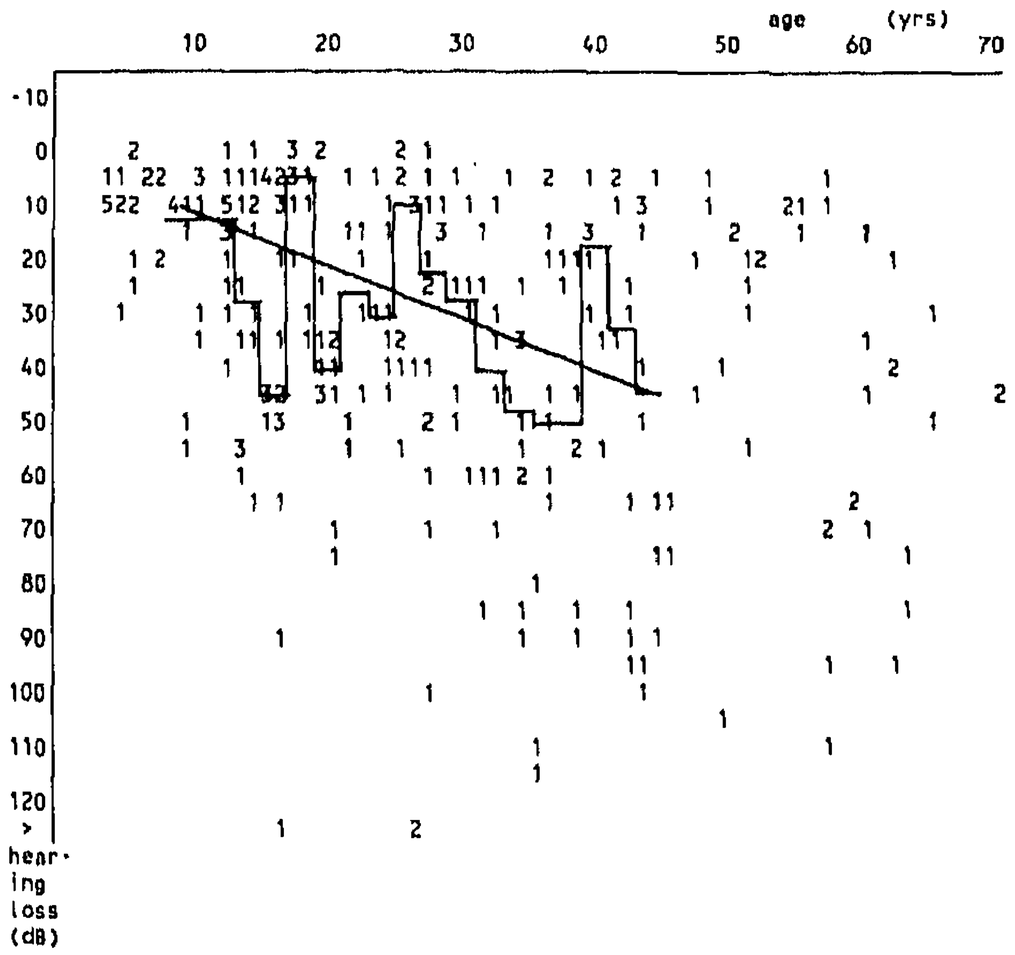

Fig 1. Mean air conduction threshold (decibels hearing level [dB HL]) for 0.5, 1, and 2 kHz (ie, Fletcher index) in selected group (282 ears, without or before surgery) plotted against age (in years). Numbers in Figure (n < 10) correspond with number of ears with given combination of certain hearing loss and age at appropriate coordinates. Histogram-like contour represents interconnected median values for hearing for age classes (width 2 years) covering 10 to 45 years of age (10-11, 12-13, ... 42-43, 44-45 years). Linear trend is shown from 10 dB at 10 years toward 45 dB at 45 years (continuous sloped line) as crude approximation to development of this "biennial" median hearing loss with age.

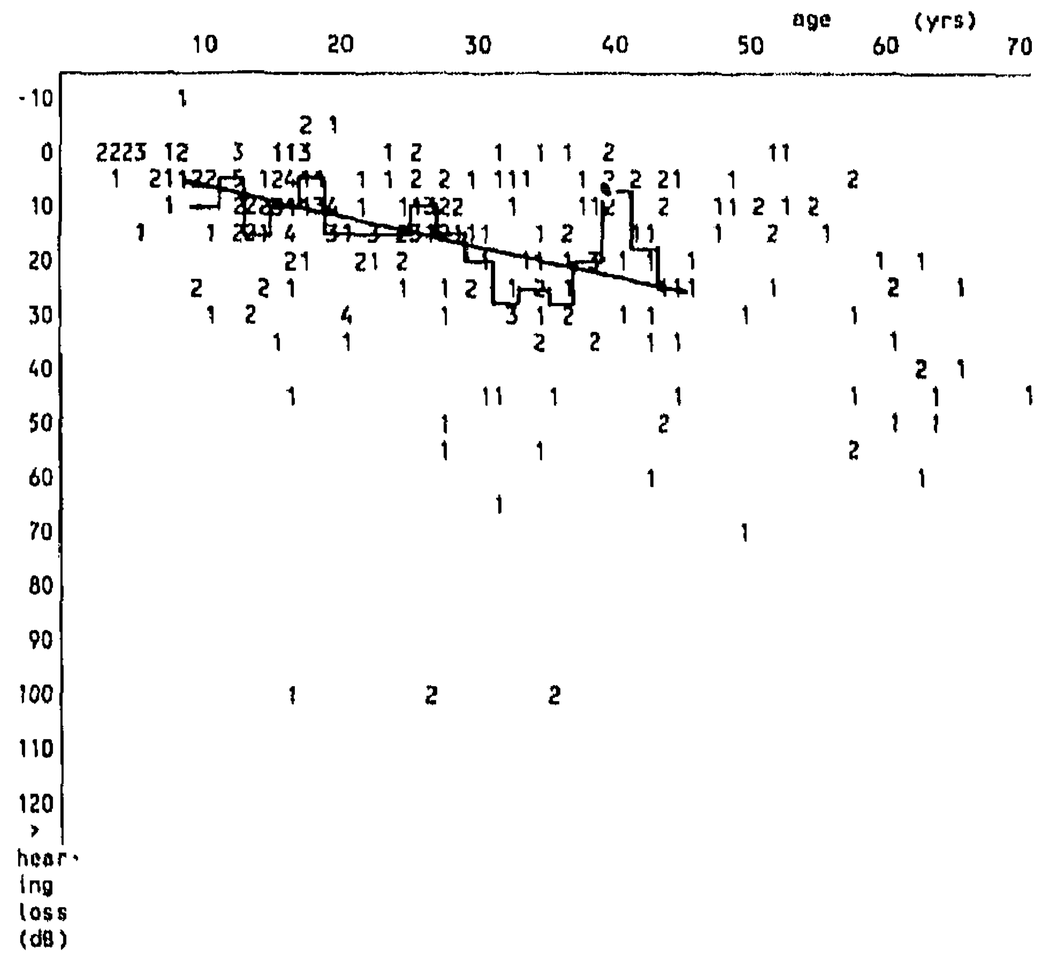

Fig 2. Mean bone conduction threshold (dB HL) for 0.5, 1, and 2 kHz in selected group (282 ears, without or before surgery) plotted against age (in years). See Fig 1 for legend. Median threshold trend is shown from 5 dB at 10 years toward 25 dB at 45 years.

hearing loss as it develops with age, we prepared separate scatter plots for the present series, for air conduction, bone conduction, and air-bone gap at various frequencies. Those for the mean hearing loss at 0.5, 1, and 2 kHz are depicted in Figs 1-3. In the plots we have indicated the median hearing loss (class width 2 years in order to obtain n > 4 for a reliable estimation of the median). It can be seen from Figs 1-3 that we utilized the median values to estimate a linear trend of hearing loss with increasing age (continuous sloped line). It should be noted that this "trend analysis" was limited to the age range 10 to 45 years for the following reasons.

1. In the age groups younger than 10 years and older than 45 years of age, there were relatively few data available, especially regarding bone conduction and air-bone gap; ie, a class width of over 2 years would be required in order to obtain n > 4.

2. This particular age range showed the most prominent increase in hearing loss.

3. The distributions of the air and bone conduction thresholds at 4 and 8 kHz, particularly in the older patients, appeared to be bimodal, analogous to those of otosclerosis[23]; this bimodality would complicate a meaningful analysis.

The results of the analysis are presented in Table 6. This Table also presents the results of a similar analysis, restricted to air conduction, in the random sample. The analysis of Table 6 yields the following results for the selected group.

1. The SNHL increased with both age and frequency. It should be noted (Fig 1) that the hearing loss (ie, air conduction) exceeded the loss that can be reasonably attributed to presbycusis, for which the mean 95th percentile threshold corresponding to the Fletcher index is about 11 dB at 20 years of age and increases through 17 dB at 45 years to about 33 dB at 70 years according to ISO 7029[22]; the 95th percentile of presbycusis appeared to coincide with the 25th percentile (averaged over 20 to 45 years) of our patient group.

2. The conductive component increased with age, to a similar degree for all frequencies.

3. The major increase in hearing loss was invariably shown by the SNHL: 0.6 dB/y for the mean of 0.5, 1, and 2 kHz; 0.7 dB/y at 4 kHz; and 1.3 dB/y at 8 kHz. The increase in air-bone gap with age was limited to about 0.4 dB/y.

DISCUSSION

The present report on a group of 142 selected subjects with OI type I shows the highest proportion of hearing loss reported so far. A comparison with the random sample[8] that formed part of this series showed that considerable selection on hearing loss had taken place at a rather young age. It is interesting

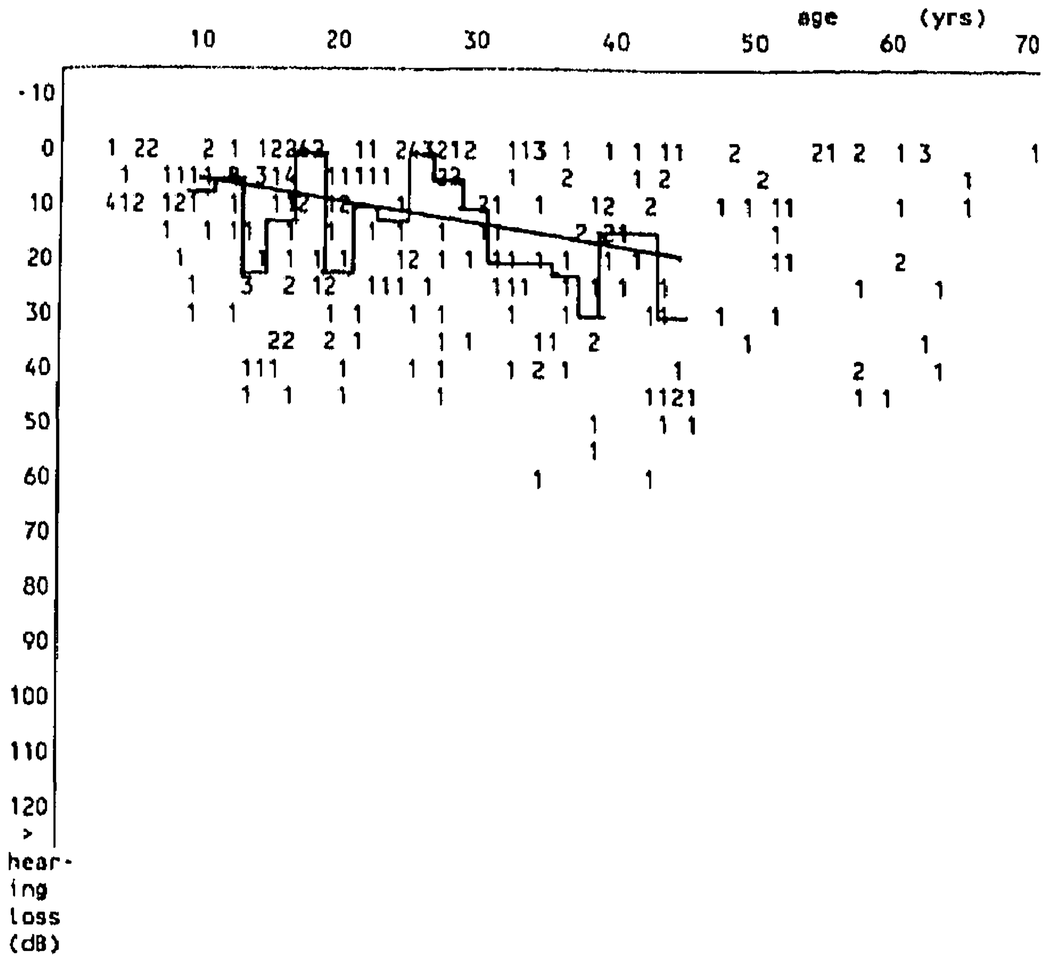

Fig 3. Mean air-bone gap (dB HL) for 0.5, 1, and 2 kHz in selected group (282 ears, without or before surgery) plotted against age (in years). See Fig 1 for legend. Median threshold trend is shown from 5 dB at 10 years toward 20 dB at 45 years.

to see that the median of the distribution of the difference in the proportion of hearing loss between the selected and the random samples — which relates to the group of affected subjects who were selected for ear surgery because of their hearing loss — coincides with the class of 30- to 50-dB loss in air conduction threshold at any age from 10 years onward. Apparently, the presumed selection has taken into account that a Fletcher index (in the nonoperated, presumably the best, ear) of more than 30 dB renders the hearing capacity less adequate socially.

The findings relating to Table 1 suggest the possibility that the family studies involved series of patients that are more similar to a random sample than to a selected sample. Therefore, it seems justified to consider the prevalence of hearing loss in OI type I as being 43%. It appears from Table 2 that the series with selected patients distinguish themselves especially by a greater hearing loss at an increasing age (30 years and older). In the present series, the proportion of subjects with hearing loss was higher than in any other series published so far, and the hearing loss occurred also at a younger age. It may be relevant that self-selection of affected subjects with hearing loss played a role, because a patient association lent its active support to the present study.

When comparing the present selected sample (Table 3) to other selected samples as closely as possible, as regards the type of hearing loss, we see that the present series included more cases of mixed hearing loss, especially in the older patients, than the series published by Shapiro et al[11] and Pedersen.[5] The present series also showed considerably more SNHL (only) at a young age as compared to those series. This only holds true as regards the comparison with Shapiro et al if CHOI is set apart. This type of hearing loss, which was found by these authors in both the lower and higher age groups in 47% of the ears, was only found in a minor percentage in the present series (Table 3), as well as in Pedersen's series. Others have also found that the predominant type of hearing impairment in OI was the mixed type, although to a much lesser degree than in the present study.[9,10,12,21] Riedner et al[9] and Stewart and O'Reilly[12] indicated that conductive loss, as far as it was present, prevailed in the younger age group. Stewart and O'Reilly, just as we did, described an age-independent proportion for SNHL.

The present series showed gradual progression in hearing loss, ie, significantly more than could be explained on the basis of presbycusis, with age: only 25% of our patients within the age range of 20 to 45 years had a Fletcher index equal to or below that derived for the 95th percentile of presbycusis. Given the large numbers of observations involved, there is

TABLE 6. MEDIAN HEARING LOSS AND ANNUAL THRESHOLD INCREASE IN OI TYPE I

		Median Hearing Loss (dB HL)			
		10 y	*45 y*	*Annual Increase (dB/y)*	
0.5, 1, 2 kHz	Bone conduction	5	25	20/35 = 0.6	
	Air-bone gap	5	20	15/35 = 0.4	
	Air conduction	10 (10)	45 (10)	35/35 = 1	(0)
4 kHz	Bone conduction	5	30	25/35 = 0.7	
	Air-bone gap	5	20	15/35 = 0.4	
	Air conduction	10 (10)	50 (20)	40/35 = 1.1	(10/35 = 0.3)
8 kHz	Bone conduction	5	50	45/35 = 1.3	
	Air-bone gap	5	20	15/35 = 0.4	
	Air conduction	10 (10)	70 (30)	60/35 = 1.7	(20/35 = 0.6)

Median hearing loss (decibels hearing level) for 142 selected patients with OI type I at ages of 10 and 45 years (estimated from linear trend), for mean at 0.5, 1, and 2 kHz, for 4 kHz, and for 8 kHz, in terms of air conduction, bone conduction, and air-bone gap, together with annual increase in threshold (decibels per year). Figures for random subsample of 70 patients with OI type I are in parentheses.

no doubt that the difference is significant. A considerable increase in hearing loss took place especially from the 10th to the 45th years of life, which comprised a considerable increase in SNHL and only a limited increase in conductive loss (Table 6). We studied individual follow-up data covering 2 through 24 years (average 9.6 years) on all subjects who had ear surgery.[15] Between 1 year and the average 9.6 years after surgery, the mean hearing level in the operated ear dropped by 6.5 dB. This almost equals the indication given in Table 6, despite the fact that the conductive component in postoperative hearing loss plays a lesser role. Also, the nonoperated ear showed a similar progression in hearing loss.[15]

It was intriguing to see that the increase in air-bone gap seemed to be limited to about 0.4 dB/y, independent of the frequency.

Less than 50% of the affected subjects older than 20 years of age had socially adequate hearing (ie, average of 30 dB or better). From a previous report,[15] it appears that among 58 operated ears, there was only 1 ear with socially adequate hearing preoperatively. Postoperatively, 24 ears (41%) did not reach the level of socially adequate hearing.

The above-stipulated progression of hearing loss with age (here evaluated in nonoperated ears only) occurs in both the nonoperated and the operated ears, and to a similar degree.[7,15] The average hearing gain immediately after the operation is about 24 dB[15]; thus, given a progression in hearing loss such as presented in Table 6, the hearing gain tends to disappear after a follow-up interval of several decades, as, indeed, seemed to be the case in the patients with a long-enough follow-up.

From an earlier study, it is clear that a relatively greater preoperative SNHL was present in the patients who were operated on at a young age.[7] It is possible that in this group of patients, the progression of SNHL that occurs preoperatively and postoperatively is greater than in the average patient. Stapes surgery is not recommended in cases of considerable hearing loss if the conductive component is too small.

Given the present findings on the natural history of hearing loss in OI, combined with our previously reported findings on the results of surgery, it seems justified to inform — if necessary — any young affected subjects whose profession depends heavily on auditory communication that they may have to cope with increasing hearing problems at a more advanced age.

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